Unicystic Ameloblastoma of the Maxilla in a 19 Year Old Patient- A Rare Case Report

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Abstract
Unicystic ameloblastoma (UA) is a benign epithelial odontogenic tumor of the jaws with an aggressive potential that commonly occurs in children. UA is not an easily diagnosable entity clinically. This cystic odontogenic neoplasm is generally asymptomatic and found during routine radiographs. The purpose of this report was to describe a rare case of UA of the maxilla in a 19-year-old patient, which was suspected to be a dentigerous cyst related to impacted premolar, and discuss its diagnosis, radiographic and microscopic findings.

Keywords: Ameloblastoma, unicystic, intraosseous, enucleation.

Introduction

Ameloblastoma as described by Robinson, is a benign tumor that is "usually unicentric, non-functional, intermittent in growth, anatomically benign and clinically persistent". Ameloblastomas are typically differentiated histologically into four chief variants- multicystic/solid, unicystic, peripheral and desmoplastic. The UA is a rare variant, which refers to those cystic lesions, that show clinical, radiographic or gross features of a jaw cyst but on histological examination shows a typical ameloblastomatous epithelium. It appears more frequently in the second or third decade with no sexual or racial predilection. The involvement of mandible to maxilla is in a ratio of 13:1.[1]

It must be differentially diagnosed form other cystic lesions in the anterior region of the maxilla, that are inflammatory radicular cysts, odontogenic keratocysts and dentigerous cysts.

Microscopically it shows a pallisaded basal layer, polarization of basal cell nuclei to the end of the cells, hyperchromatism of the basal cells, vacuolation of the basal cell cytoplasm, stratum spinosum like stellate reticulum and sometimes keratinization.

Portions of UA showing ameloblastic epithelium and secondary inflammation make the diagnosis very difficult. Importantly, ameloblastic epithelium must not penetrate the wall of the cyst. If so, the lesion behaves as a solid ameloblastoma. Microscopic variants include luminal type in which the ameloblastic epithelium is confined to the luminal layer and the intraluminal in which nodules of ameloblastic epithelium project into the cyst lumen. These lesions are destructive and 15% may recur after simple enucleation.

Here we report a rare case of unicystic ameloblastoma of the maxilla in a 19-year-old male.

Case report

A 19-year-old boy reported to our clinic with a swelling on the right side of the cheek. The swelling was slowly growing over a period of one year and was not associated with pain, tenderness or discharge. Past medical history and dental history were unremarkable. He had no known drug allergy and was not on any medication. His general physical examination revealed no other abnormality. On local extra oral examination, the swelling extended diffusely from infraorbital margin superiorly to the angle of mouth inferiorly and from the zygomatic buttress laterally to the nasolabial fold medially, causing slight distortion of the face (Figure 1). The overlying skin had normal colour and temperature. On palpation, it was bony hard and non tender. Intraoral examination showed a swelling on the buccal aspect, extending from the right maxillary canine to the first molar. The deciduous first molar was retained and first premolar was missing from the first quadrant. The overlying mucosa was smooth and there was no colour change (Figure 2). The OPG showed a well-defined unilocular radiolucency extending from 12 to 17 anteroposteriorly, and from infra-orbital margin to the alveolar crest superio-inferiorly, which was associated with retained 54 having resorbed roots, impacted 14 and displaced 16 and 17 (Figure 3).

On aspiration, a yellow straw-colored fluid was obtained and provisional diagnosis of dentigerous cyst in relation to impacted 14 was made. The lesion was enucleated (Figure 4), 54 and 14 removed, along with intranasal antrostomy. Grossly, the greyish white soft tissue specimen was around 4x3cm in dimensions and soft in consistency. On microscopic examination, the cystic space was lined by an epithelium lining which demonstrated a basal layer of ameloblast like cells, showing pallisading, reverse polarity and sub-nucleolar vacuolization. The overlying cells were star shaped and loosely cohesive, resembling the stellate reticulum. The lesional cells showed intraluminal as well as mural proliferation (Figure 5,6). The final histopathological diagnosis was unicystic ameloblastoma**.** The patient was followed up for 2 years with no evidence of recurrence.

Discussion

Unicystic ameloblastoma, a variant of ameloblastoma, was first described by Robinson and Martinez in 1977. The unicystic ameloblastoma is more correctly referred to as a mural ameloblastoma in situ. A study of 21 cases of UA by

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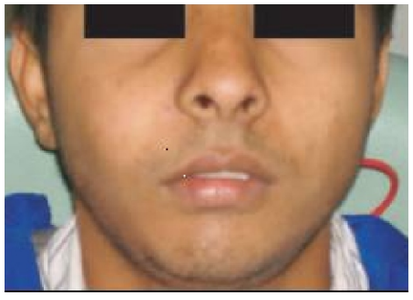

Figure 1: Extraoral view showing a swelling on the right side middle third of face

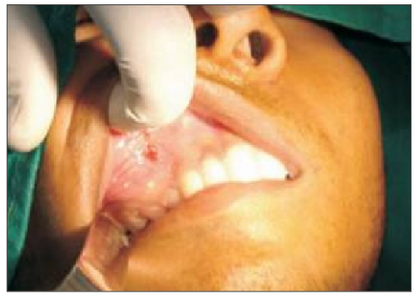

Figure 2: Intraoral view- swelling extending from 13 to 16 with nornal overlying mucosa

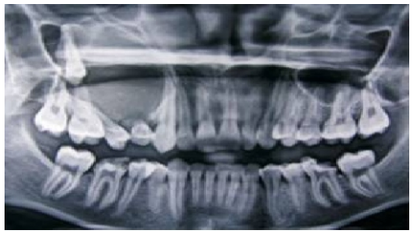

Figure 3: OPG showing a well defined unilocular radiolucency in the right maxilla

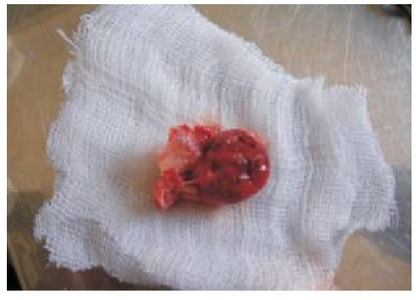

Figure 4: The enucleated specimen

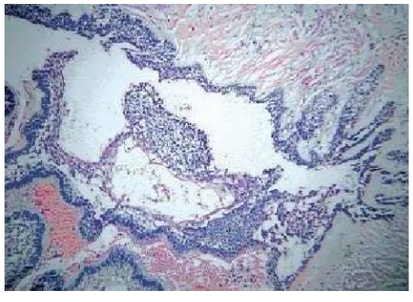

Figure 5: Low power photomicrograph shows ameloblastomatous cystic epithelial lining, having a luminal, intraluminal, as well as mural proliferation (H & E stain, 10X)

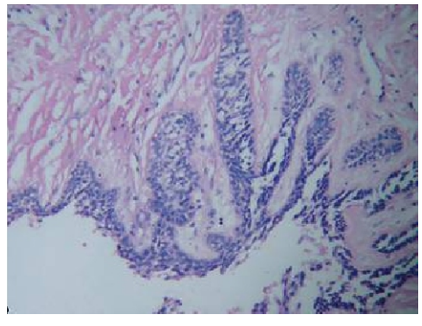

Figure 6: Photomicrograph shows a high power view of the same lining epithelium (H & E stain, 20 X)

Rosentein et al., revealed that the age range of patients with cystic ameloblastomas was 12 to 72 years, with a mean of 35 years, with almost equal gender distribution. However all the 21 cases in this study were found in the mandible.[2] Similarly, other studies done by Gardner and Corio, by Robinsen and Martinez and by Eversole, Leider and Strub also found that all their cases of UA were present in the mandible.[3]

UA is a variant of the solid or multicystic ameloblastoma. Radiographically, the unilocular pattern is more common than the multilocular, especially in cases associated with tooth impaction. However, it is stressed that although the lesion is pathomorphologically unicystic, it will far from always produce a unilocular radiolucency. The mean age at the time of diagnosis of UA is closely related to its association with an impacted tooth. There is a difference of about 20 years in the mean age of the 'dentigerous' variant from the 'non-dentigerous' (16.5 years versus 35.2 years). The male:female ratio for the 'dentigerous' type is 1.5:1, but for the 'non-dentigerous' type it is reversed (1:1.8). Location favours greatly the mandible (mandible:maxilla = 3 to 13:1).[4] Between 50% and 80% of cases are associated with tooth impaction, the mandibular third molar being most often involved. The 'dentigerous' type occurs on average 8 years earlier than the 'non-dentigerious' variant. The mean age for unilocular, impaction-associated UA's is 22 years, whereas the mean age for the multilocular lesion unrelated to an impacted tooth is 33 years.[4]

One study revealed an interesting finding about these lesions, that the mean age for non impacted tooth-related cystic ameloblastomas was 35 years in comparison to 16.5 years for the impacted tooth-related (dentigerous) variant,[5] and is consistent with our case, where the patient's age was 19 years and was associated with an impacted premolar.

The location of this lesion in the anterior region of the maxilla is considered to be rare and atypical, since this lesion predominantly occurs in the mandible, with the molar region and the ascending ramus being the most affected areas. The ratio of mandibular to maxillary unicystic ameloblastoma has been reported to be 13:1. Similar location has been reported in a 17 year old white male by Navarro et al., in 2004.[6]

Differential diagnosis for this tumor in this particular location includes radicular cyst, odontogenic keratocyst, ameloblastoma, adenomatoid odontogenic tumor, ameloblastic fibroma, odontogenic myxoma, and glandular odontogenic cyst. The definite diagnosis cannot be ruled out on clinical or radiographic grounds; histopathologic confirmation is required for diagnosis.[7]

Ackermann classified this entity into the following three histological groups:

GroupI: Luminal UA (tumor confined to the luminal surface of the cyst)

GroupII: Intraluminal/plexiform UA (nodular proliferation into the lumen without infiltration of tumor cells into the connective tissue wall), and

GroupIII: Mural UA(invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium).[7,8]

Another histological subgrouping by Philipsen and Reichart has also been described:

Subgroup 1: Luminal UA

Subgroup 1.2: Luminal and intraluminal

Subgroup 1.2.3: Luminal, intraluminal and intramural

Subgroup 1.3: Luminal and intramural

The UA s diagnosed as subgroups 1 and 1.2 can be treated conservatively (careful enucleation), whereas subgroups 1.2.3 and 1.3 showing intramural growths require radical resection, as for a solid or multicystic ameloblastoma. Following enucleation, vigorous curettage of the bone should be avoided as it may implant foci of ameloblastoma deeper into bone. Chemical cauterization with Carnoy's solution is also advocated for subgroups 1 and 1.2. Subgroups 1.2.3 and 1.3 have a high risk for recurrence, requiring more aggressive surgical procedures.[7]

Pathogenesis of this lesion remains obscure. Why some ameloblastomas become completely cystic may be related to epithelial dysadhesion (e.g., defective desmosomes), or more likely to the intrinsic production of proteinases (e.g., metalloproteinases or serine proteinases); enzymes that normally degrade the central portion of enamel organ after tooth development.[3]

Recurrence rate of 15% has been reported after enucleation and curettage of unicystic ameloblastoma. This is considerably less than 50-90% recurrence rate noted after curettage of conventional solid ameloblastomas.[9-12] UA is difficult to diagnose. It shows considerable similarities with dentigerous cysts. It has been proposed that UA may take its origin from a pre-existing dentigerous cyst. The lining of UA is often lined partly by a nonspecific thin epithelium that mimics the dentigerous cyst lining.[13] Histological examination may be the only means of differentiating between the two.

If UA is limited to the epithelial lining of a cyst-like lesion, simple enucleation can be performed as was done in our case. The young age of the patient is also a criterion while determining the treatment to be performed. We prefer to go in for a therapy which has minimum surgical trauma, least restriction of jaw function and leads to minimum jaw deformation. Hence, a simple enucleation was performed in our case. However, if epithelial cells extend and proliferate into the fibrous tissue surrounding the epithelium, it must be regarded as an invasive lesion and wide excision/ resection may be considered in conjunction with other clinical and pathological factors such as the size, location and growth pattern of the tumor. Recurrence of unicystic ameloblastoma may be long delayed and a long-term postoperative follow up is essential for the proper management of such patients.[14]

Summary

The diagnosis of unicystic ameloblastoma is based on clinical, radiographical and histopathological features. The simple subtype with and without intraluminal proliferations may be treated conservatively (enucleation), whereas subtypes showing intramural growths must be treated radically, i.e, as a solid or multicystic ameloblastoma.

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